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Parapharyngeal lymphangioma -a case report

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Abstract: *Parapharyngeal lymphangioma means cystic hygroma involving parapharyngeal region which can obstructs the airway laterally, this is very rare condition. We present here a case report of a 9-year-old femalepatient who came with a chief complaint ofpain and swelling on left side of face since 8 days and was later diagnosed as having cystic hygromain parapharyngeal region on the basis of her clinicalfeatures and further imaging studies.This report also highlights the surgical and other treatment options.*

Keywords: *cystic hygroma, lymphangioma, parapharyngeal Lymphangioma, swellings in neck, sclerotherapy.*

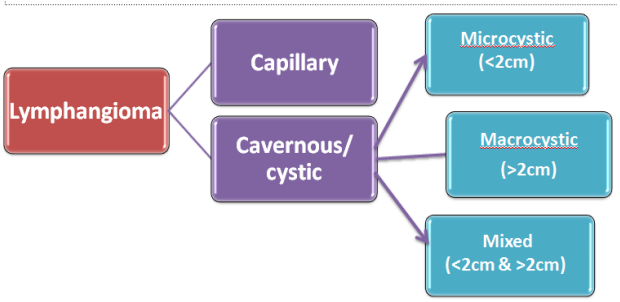
I. Introduction

Cystic Hygroma was first described in the European Literature by Redenbacher in 1828[1].Cystic hygroma is also known as cystic Lymphangioma, jugular lymphatic obstructive sequence, hygromacollicysticum. The term hygroma means moist tumor. Lymphangiomas are usually classified as shown in fig.1. Cystic hygroma are anomalies of the lymphatic system characterized by single or multiple cysts within the soft tissue, usually involving the cervico-facial regions and axilla. Other less common sites are mediastinum, groin and below tongue. Occasionally, these malformations occur in liver, spleen, kidney and intestine. Omental cyst in omentum and mesenteric cyst in the mesentery of intestine represents parallel lesions at these locations[4].Respiratory distress, recurrent infections or cosmetic reasons are the main indications of the treatment[2].Exact etiology is unknown.

There are a number of proposed mechanisms to explain the pathophysiology of cystic hygroma. Embryologically, these lesions are believed to originate from sequestration of lymphatic tissue from lymphatic sacs, during the development of lymphatico-venous sacs. These sequestered tissues fail to communicate with remainder of the lymphatic or venous system. Later on, dilatation of the sequestered lymphatic tissues ensues, resulting in the cystic morphology of these lesions[2].They occur most commonly in the neck, which is termed nuchal cystic hygroma (occurs in ~80% of cases) and axilla, with only 10% of cases extending to the mediastinum and only 1% confined to the chest[13]. The incidence of cystic hygroma is not well defined. Reports range from 1.7:10,000 pregnancies to 0.83% of pregnancies at risk for having a structural anomaly[5] and there is no hereditary predisposition exists. Over 50% of cystic hygroma present at birth and more than 90% of these congenital malformations are found in children younger than 2 years with males and females equally affected. They usually occur inthe fetal/infantile and paediatricpopulations with most lesions presenting bythe age of two. The estimated prevalence in the fetal population is 0.2-3%. It is quite rare in adult[4,5].

They occur most commonly in the neck, which is then also termed nuchal cystic hygroma (occursin ~80% of cases) and axilla, with only 10% of cases extending to the mediastinum and only1%confined to the chest[13].The usual presentation of cystic hygroma apparent at birth is a painless mass with worries and queries of the parents about the lesion. The other modes of presentations are related to the complications or effects of cystic hygroma, such as respiratory distress, feeding difficulty, fever, sudden increase in the size and infection in the lesion.[14,15] On clinical examinations, these lesions appear soft, compressible, non-tender, transluminant and without any bruit. Intraoral examination reveals bulging in posterior region of soft palate or pharyngeal wall. In case of lesion anterior lower neck mass causes elevation of tongue, floor of mouth and inability to close the mouth. The other modes of presentations are related to the complications or effects of cystic hygroma, such as respiratory distress, feeding difficulty, fever, sudden increase in the size and infection in the lesion.

Differential diagnosis reveals dentoalveolarabcess, mumps, sialadenitis, Sjogren's syndrome, benign and malignant tumors of parotid gland.



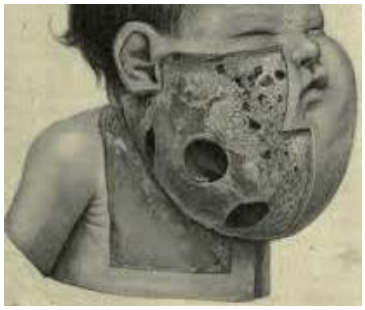


fig.1: classification of lymphangioma **fig. 2:**multicystic appearance of lymphangioma.

II. Case Report

A 9 year old female Hindu patient presented with a swelling on left side of the face which was gradually increasing in size since 8 days, she also developed difficulty in breathing and pain on palpation. History of present illness reveals patient had same swelling two years back for that she had taken antibiotics and anti-inflammatory medication. Then the swelling get subsided and recurred 3-4 times up till now. Physical examination revealed a 8 X 6cm bulging soft mass over the left side of the neck, extending 3cm anterior to the ala tragus line superio- anteriorly to the 2cm posterior to ear superio-posteriorly. Inferiorly it involves midline of mandible at anterior end to angle of mandible at posterior side giving an oblique elongated appearance (Fig. 3 a and b). Theswelling was a soft, compressible, elongatedand tender on palpation. Local temperature was normal. Thetransillumination testwas positive.Small other firm tender masses were palpable surrounding the biglobulated mass. The patient's voice was normal.There was no cervical or supraclavicularadenopathy.Initially 7 days antibiotic course was given.Intraoral examination showed bulging of left faucialpillar, soft palate and pharyngeal wall.

Prior sonography report suggested the presence of salivary gland inflammation. RecentPulsed Doppler sonography image of the neckposterior to the left mandibular angle shows large well defined multisepted cystic mass lesion involving parapharyngeal, lingual and parotid space. Measuring 8×6cm in size consist of thick fluid inside the cystic cavity. Absence of any calcification noted.The lesion is seen adjacent to tonsillarfossa and tongue; this also displaces blood vessels in neck postero-laterally. No movement observed withtongue movement and deglutition. These findings denotelymphocele or cystic hygroma.

The echogenic portions of the lesion correlate with clusters of small, abnormal lymphatic channels. Fluid filled level can be observed with a characteristic echogenic component layering in the dependent portion of the lesion.

CECT scan of the neck and thorax showed a large loculated cystic mass lesion of nearwater density with few enhancing septa within. It occupied the posterior cervical space on the left side of the neck lateral to the carotid artery. Axial postcontrast CT image of the neck at the level of the mandibular angle obtained during the current admissionit displaced the left internal jugular vein anterio-medially and the sternocleidomastoid muscle interio-laterally. In its cranio-caudal extent, it extended from C1 to the level of vertebra C4.

Axial postcontrast CT image of the neck obtained reveals a largewell defined unilateralmultiloculatedhypodense lesion more prominent over the left submandibular region spreading through the soft tissue measuring about 8×6×6cm in size. The major component of the lesion involved the left side of the neck from the level of the parotid gland to the thoracic inlet.There was also moderate enlargementof the adenoids and tonsils, which, together with theparapharyngeal space was partially obstructing the airway.Report of routine blood, urine and x-ray chest examination was normal.

After FNAC, microscopic examination comprised of endothelium lined cystic spaces with scanty stroma. A final diagnosis of cystic hygroma was made.

Because of the increasing size and pain during thecurrent admission, it was elected to remove the lesionssurgically. Transcervical transmandibular approach through the digastric triangle wasused to access the submandibular and parapharyngeal space. Submandibular gland excision done for good exposure. Multiple lymphfilled sacs wereencountered. A cystic area with old hemorrhagicelements due to prior bleeding was observed in the leftside of the neck posterolateral to the mandibular angle.Complete excision of mass removed in toto. Neck drain kept rest postoperative period was uneventful.

Histopathology revealed thin connective-tissue stromaseparating the cystically dilated spaces lined by asingle layer of benign endothelial cells which wasconsistent with cystic hygroma.The patienttolerated the procedure well and suffered only minorweakness of the marginal mandibular branch of the facialnerve.

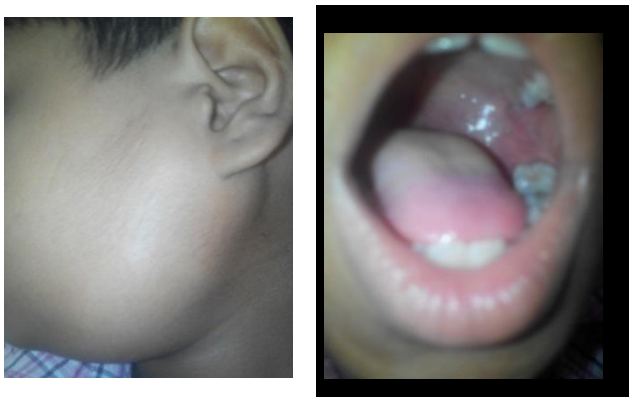


Fig. 2 (a) Extraoral and(b) intraoral appearance of lesion.

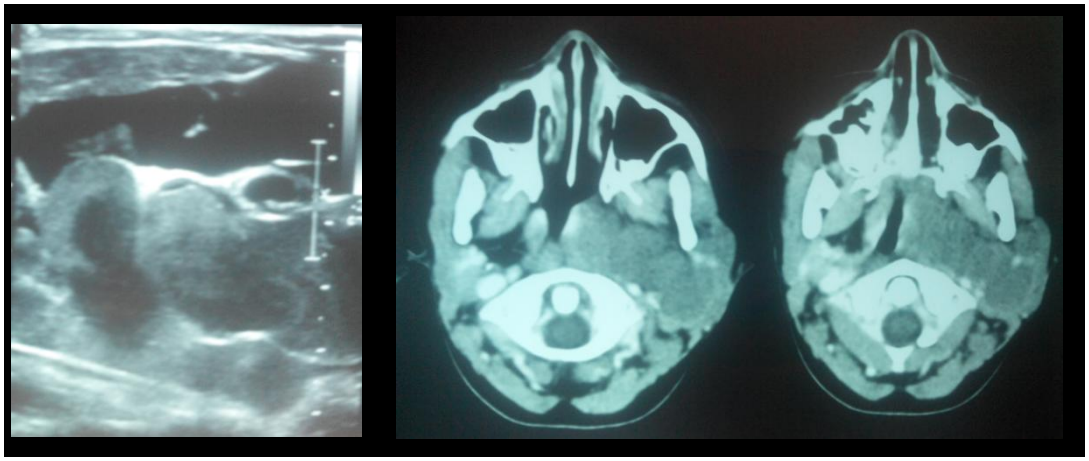


fig.3 : ultrasound image shows multicystic appearance.

fig.4: axial CT image shows parapharyngealextention of lesion obstructing the airway.

fig. 5: excised cystic mass of 6x8 cm size.

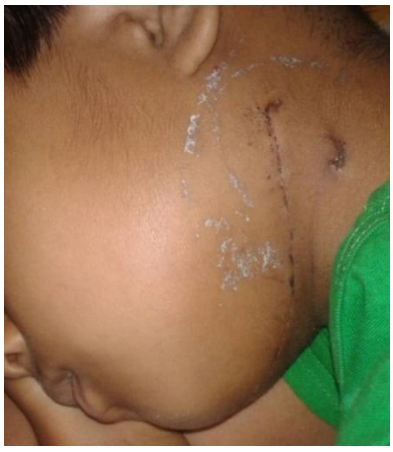


fig. 6: one month after surgery.

III. Discussion

Cystic hygromas are congenital vasculolymphaticmalformations that are frequently present at birth. Theyhave no predilection for sex or race, and they have nomalignant potential. Typical cystic hygromas cause nosymptoms unless they enlarge in size or surround orinvade adjacent normal anatomic structures. In thissituation, cystic hygromas may cause symptoms such asfeeding problems or breathing difficulties[2]. Our patient showed a tender swelling with a little difficulty in swallowing.

They usually occur in the fetal/infantile and paediatric populations with most lesions presenting bythe age of two.The estimated prevalence in the fetal population is 0.2-3%.Patients in the infantile or paediatric population can present with pain, dyspnoea, infection,haemorrhage or respiratory compromise. In contrast our patient was 9 yr old and she never felt any swelling or respiratory discomfort before.

Pathologically, Cystic hygromas are multilobulated, thin-wall, lymphcontaining sacs. The fluid within the sacs is usually produce milky, serous, sero-sanguinous or strawcoloured fluid, when aspirated with a widebore needle[4]. Cystic hygroma are thought to arise from delayed development/maldevelopment/failure of the lymphaticsystem to communicate with the venous system of the neck. Like other lymphangiomas, they are endothelial lined cavernous lymphatic spaces.

Theycan vary significantly in size. Lymphatic vascular malformations may be mixed with other formsof vascular malformation, including capillary or venous.

They occur most commonly in the neck, which is then also termed nuchal cystic hygroma (occurs in ~80% of cases)[7] and axilla, with only 10% of cases extending to the mediastinum and only1% confined to the chest. In our case the lesion was above neck region and deep involvingparapharyngeal regionupto airway and less likely to bulging peripherally[5].

Associated with aneuploidic anomalies: ~65% (range 50-80%) like Turner syndrome : Down syndrome, trisomy 13, trisomy 18, triploidyand non aneuploidic like congenital cardiac anomaliesaortic coarctationhypoplastic left heart syndrome, pentalogy of Cantrell, Apert syndrome, Cornelia de Lange syndrome, fetal alcohol syndrome, Fryns syndrome, lethal multiple pterygium syndrome, limb hypertrophy, Noonan syndrome, Pena Shokeir syndrome.[5,14]

The mainstay of lymphatic malformation treatment has been surgical resection, which has been refined through lesion and radiographic characterization.Sometimes, this may be impossible due to the infiltrating nature of the hygroma within and around neurovascular structures, muscles and blood vessels.[16]

In this condition, unroofing, partial cystectomy anddrainage of the cystic content should be performed andall adjacent crucial structures should be preserved. Recurrence rate of 10-15 % isreported.[10,11]

The other techniques recently introduced in the management of lymphangiomas are radiofrequency ablation and laser excision(using CO_2, Nd-YAG, or potassium-titanylphosphate lasers) is minimally invasive and is much less painful than surgery.[13] Complications following resection arepostoperative muscle weakness, nerve injuries andneural weakness, bleeding and wound infection. In ourpatient the spinal accessory nerve was splayedanteriorly all across over the cystic mass. On themedial aspect, the mass could be carefully separatedfrom the facial nerve, parotid gland, internal jugular vein, carotid vessels, vagusnerve and hypoglossal nerve.

An alternative to surgery, intralesionalsclerotherapy in macrocystic lymphatic malformation, is effective and reduces the need for other forms of therapy for some cases.Intralesionalsclerosing agent like bleomycin, OK-432, boiled water, quinine, sodium morrhuate, urethane, iodine tenture, doxycycline and nitromin are used after aspirating the cystic fluid[17].

IV. Conclusion

The cystic hygromas situated in parapharyngeal region calls for extra attention as it is deep neck spaces and containing important neurovascular structure and per se lymphangioma of parapharyngeal space is rare entity. It is difficult to diagnose it purely on clinical examination .Preoperative imaging plays crucial role in identification of exact origin such lymphatic anomalies. This information can help to delineate the vascular surgical map and avoid complications during surgical exploration. The use of reconstructed CT images and color Doppler sonography are useful to evaluate the neck lesions and analyze their relationship to adjacent anatomic structures. Transcervicaltransmandibular approach provides good exposure of parapharyngeal space and for excision such lesion. Considering rarity of entity cystic hygroma of deep neck space will be interesting reading for all.

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